Understanding the Complications of Sickle Cell Disease

An evidence-based review of essential nursing assessments, teaching points, and areas of advocacy.

ABSTRACT: Sickle cell disease (SCD) is an autosomal recessive genetic condition that alters the shape and function of the hemoglobin molecule in red blood cells. While the overall survival rate among children with SCD has improved in recent years, pediatric rates of hospitalization, ED use, and mortality from complications of SCD remain high. Among patients ages 18 and older, hospital admission and ED usage are even greater—and the median age at death of people with SCD is considerably lower than that of the general population. Nurses who care for patients with SCD have an opportunity to improve health outcomes and quality of life for these patients by recognizing the major SCD-associated complications and providing patients and their caregivers with appropriate educational information. The authors discuss the genetic, hematologic, and clinical features of SCD and describe the major associated health complications. In addition, they review the nursing implications of each complication and provide online links to resources for clinicians, patients, and caregivers.

Keywords: sickle cell anemia, sickle cell disease, hemoglobin, hemoglobinopathy

Sickle cell disease (SCD) is an autosomal recessive genetic condition that alters the shape and function of the hemoglobin (Hb) molecule, causing red blood cells to take on the shape of a sickle (or crescent) (see Figure 1). The sickled blood cells break down prematurely, potentially producing anemia. Since they are rigid, they may become trapped in small blood vessels, triggering acute painful events, depriving tissues of oxygen-rich blood, and damaging organs, most notably, the spleen. Splenic injury greatly increases the risk of death from infection at a young age.

Millions of people throughout the world have SCD, most frequently those of African, South or Central American, Caribbean, Mediterranean, Indian, or Saudi Arabian descent. In the United States, it is the most common inherited blood disorder, affecting more than 100,000 people, approximately 90% of whom are black and 10% of whom are Hispanic.[1] (The number of non-black or non-Hispanic SCD patients is too low to estimate based on currently available data.) Of the four primary SCD genotypes, HbSS and HbSβ^0-thalassemia tend to be the most clinically severe forms of the disease and are collectively termed sickle cell anemia (SCA). Generally, HbSC and HbSβ^+-thalassemia are the more clinically benign forms, though severity varies among patients, and even patients with these typically milder forms of SCD commonly experience acute painful events and such serious complications as acute chest syndrome (ACS), silent cerebral infarction (SCI), avascular necrosis (AVN), priapism, and kidney failure throughout life.[2-6]

In 1987, the National Institutes of Health (NIH) convened a Consensus Development Conference on

Newborn Screening for Sickle Cell Disease and Other Hemoglobinopathies, which recommended that all newborns in the United States be screened for SCD.[7] It was not until 2006, however, that all 50 states and the District of Columbia had fully adopted the recommendation.[8] Now, the identification of SCD in newborns enables physicians to initiate prophylactic penicillin and early vaccination against *Streptococcus pneumoniae* in order to prevent related infection or death, and provides family members an opportunity to learn how to reduce the child's risk of potential SCD-related complications, to consider the implications of the child's condition for their own and the child's future family planning, and to seek evidence-based care for the child from health care professionals with an expertise in SCD. A 2010 study of the Dallas Newborn Cohort, which includes children who were diagnosed with SCD at birth (through newborn screening) and received ongoing care at a Dallas-based, tertiary SCD clinic from 1983 forward, found that the overall survival rate at age 18 was nearly 94% among children with SCA (up from 86% in 2004) and 99% among those with the less severe HbSC or HbSβ$^+$-thalassemia (up from 97% in 2004). ACS and multisystem organ failure had replaced bacterial sepsis among these children as the leading causes of death.[9]

Despite increased survival among pediatric patients with SCD, hospitalization and ED usage rates are estimated to be seven to 30 and two to six times higher, respectively, than those of same-age cohorts without the disease.[10] ED visits, hospital admissions, and hospital readmissions are even more frequent among 18-to-30-year-olds with SCD than among children or adults in middle or older age.[11] The median age at death among patients with SCD is considerably lower than that of the general population: for men and women with SCA, 42 and 48 years, respectively; for men and women with HbSC, 60 and 68 years, respectively.[12]

Two of the greatest challenges faced by clinicians caring for patients with SCD are the lack of evidence-based guidelines, owing to the paucity of data from large, randomized controlled trials involving SCD patients, and the underuse of the few recognized disease-modifying therapies.[13] Since, in the United States, SCD affects primarily black and Hispanic patients, it's been suggested that "conscious or unconscious racial bias" has limited the availability of resources for research, as well as for the delivery and improvement of care.[14] An evidence-based report on the management of SCD, supported by the NIH and published by the National Heart, Lung, and Blood Institute (NHLBI), identified only seven areas in which strong recommendations based on high-quality evidence could be made.[13,15] (See *NHLBI Expert Panel Recommendations for Managing Sickle Cell Disease*.[13,15]) The other recommendations are based on observation or low-quality evidence. This article reviews the major health complications faced by patients with SCD, discusses the nursing actions relevant to each, and provides a summary of these for quick reference (see Table 1). It also lists online links to several resources for clinicians, patients, and patient caregivers (see *Sickle Cell Disease Clinical Care Resources*), as well as little-known facts about SCD (see *Facts About Sickle Cell Disease*[5,9,10,16-19]).

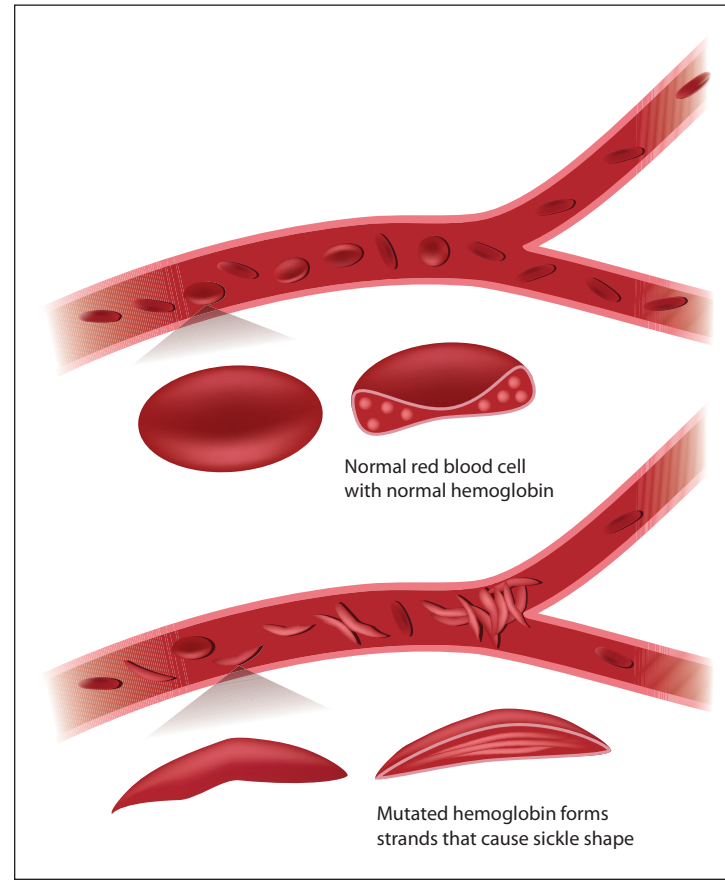

Figure 1. Normal red blood cells are round and flexible and move easily through blood vessels. In sickle cell disease, abnormal hemoglobin causes red blood cells to become sickle (or crescent) shaped and rigid. The misshapen cells can easily become lodged in smaller blood vessels, depriving tissues of oxygen and triggering painful episodes. Illustration by Alila Medical Media.

ACUTE PAIN

An acute vasoocclusive episode (VOE) is a new onset of severe pain that persists for at least four hours

for which there is no explanation other than vasoocclusion. VOEs are the primary source of acute pain requiring medical attention in SCD, and they account for most acute pain episodes experienced by patients with this condition. VOEs can be treated with parenteral opioids, ketorolac, or other nonsteroidal antiinflammatory drugs. As patients grow older and learn strategies for alleviating the pain of VOEs, they often manage these episodes at home, but VOEs are the most common manifestation of SCD and the most common SCD-related ED diagnosis.[16] In a 2013 follow-up analysis of 264 adult patients with SCA in the Bethesda Sickle Cell Cohort study, approximately 40% reported no hospitalizations or ED treatments for VOEs within the past year, whereas 22% reported five or more hospitalizations or ED visits within that period, underscoring the tremendous variability of VOE severity that occurs in SCA.[20]

The clinical management of VOEs focuses on rapid pain assessment and administration of appropriate analgesics. Infants and toddlers often present with dactylitis, or pain and swelling from infarctions in the hands and feet; these may be the first SCD-related complications they experience (see Figure 2).[21] Other common locations for VOE pain in patients of all ages include the chest, abdomen, back, and long bones. SCD genotype, older age, higher hematocrit levels, and lower fetal Hb levels are risk factors for VOE.[12] Patients may have specific triggers for pain, such as extremes in temperature, dehydration, menstrual cycle changes, alcohol, or stress, but the majority of painful episodes have no identifiable cause. The effects of VOEs extend beyond the episodes themselves. Acute multisystem organ failure and death have also been reported during VOEs.[22,23] And patients with more frequent VOEs are shown to have a younger age at death.[20]

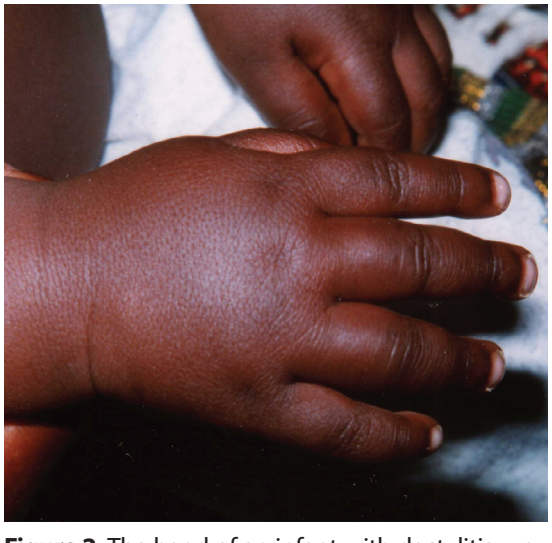

Figure 2. The hand of an infant with dactylitis—a painful swelling of the fingers or toes. Photo courtesy of Tom D. Thacher, MD.

Nursing implications. A nurse's comprehensive pain assessment, noting location, intensity, and duration of the pain episode, is invaluable. Nurses help patients identify what normally relieves the pain and whether the patient's current pain is a typical VOE or suggests a different complication, such as ACS. As primary advocate for the patient, nurses can work with the prescribing clinician to ensure the patient receives adequate analgesia. Nurses can also work with patients to determine which nonpharmacologic adjunct therapies best relieve the pain. Most patients find heat application comforting; the application of ice is contraindicated in VOEs because vasoconstriction increases the pain by reducing oxygen delivery. Other nonpharmacologic therapies frequently used in SCD include cognitive behavioral therapy, biofeedback, prayer, relaxation techniques, acupuncture, hypnosis, herbal therapies, and megavitamins.[24]

After the pain has been relieved, nurses can work with patients to understand potential VOE triggers and how to avoid them and to ensure patients understand how to take the pain medicines prescribed to manage their pain. Nurses may also help patients understand the importance of hydration in preventing VOEs by increasing plasma volume and reducing blood viscosity. Unless contraindicated by conditions such as heart failure or renal disease, fluid intake may be encouraged. Finally, nurses can persuade patients, who may be reluctant to move because of pain, to walk and use the incentive spirometer every hour to prevent blood clots and pulmonary complications.

CHRONIC PAIN

Three subtypes of chronic pain have been identified in SCD[25]:

- chronic pain without contributing SCD complications, such as leg ulcers or AVN
- chronic pain with contributing SCD complications
- mixed presentation, when there is evidence of chronic pain from SCD complications but also apparently unrelated persistent pain

As with other types of chronic pain, SCD pain can cause patients to experience central sensitization, hyperalgesia, and altered opioid metabolism. Chronic pain not only causes discomfort but may also precipitate mood changes, emotional disturbance, and behavioral dysfunction, thereby affecting numerous facets of the patient's life as well as the lives of family members, friends, and colleagues.[26] In a six-month, prospective cohort study of 232 patients with SCD who were age 16 or older, 29% experienced pain almost daily while 14% experienced pain on 5% or fewer days.[27] This chronic pain was more frequent than VOEs and was often managed outside of a health care setting.

Nursing implications. In addition to conducting a comprehensive pain assessment for acute VOEs,

Table 1. Major Complications of Sickle Cell Disease and Nursing Implications

Complication	Nursing Implications
Acute pain from VOEs	• Conduct a comprehensive pain assessment. • Advocate for appropriate pain management. • Help patients understand potential triggers and avoidance strategies. • Ensure patients understand how to take pain medicines to manage acute pain. • Encourage fluid intake (unless contraindicated, as in the presence of heart failure or kidney disease), ambulation, and incentive spirometry.
Chronic pain	• Understand the SCD complications that can contribute to chronic pain. • Perform a comprehensive patient assessment and history. • Obtain a thorough medication history. • Teach patients which types of chronic pain are amenable to different interventions and how to use both medications and nonpharmacologic interventions most effectively.
AVN	• Conduct a thorough chronic pain assessment (type of pain and underlying mechanism) and maintain a high index of suspicion for AVN. • Refer patients experiencing hip pain for orthopedic consultation.
Priapism	• Teach patients that priapism is a potential complication of SCD. • Emphasize the importance of reporting events to prevent adverse sequelae and of seeking medical attention for prolonged episodes within 4 hours of onset. • Answer patient questions with the understanding that the topic of priapism can be uncomfortable and anxiety inducing. • Remind patients who experience priapism to pay attention to precipitating factors. • Be sensitive to the potential psychological effects of priapism on patients.
ACS	• Conduct a comprehensive respiratory assessment, noting even subtle changes in respiratory status. • Teach and encourage the use of incentive spirometry. • Report any changes in respiratory status to the primary care provider.
Stroke	• Conduct neurologic assessments routinely in children and adults and maintain a high index of suspicion for SCI in patients who demonstrate neurologic deficits. • Assess parents' understanding of the need to seek care for any emerging neurologic symptoms. • Teach parents about the importance of routine TCD ultrasound screening in children with SCA. • Remind parents that a child's poor academic performance may signal neurocognitive deficits resulting from SCIs. • Encourage parents to discuss poor academic performance with the health care team. • Report any acute changes in neurologic status to the primary care provider.
Splenic complications	• Conduct a thorough assessment of abdominal pain, closely monitor temperature, and anticipate the need for a sepsis evaluation. • Teach parents the importance of immunizations and of recognizing fever early and notifying the child's health care team immediately.
Infection and sepsis	• Monitor vital signs and report elevations in temperature to the primary care provider. • Teach patients or their parents the importance of monitoring fever and receiving age-appropriate immunizations.
Organ failure	• Monitor renal function and IV fluid administration, especially in patients with a history of renal failure. • Assess kidney disease risk factors throughout hospitalization and maintain a high index of suspicion for proteinuria or reduced urine output. • For patients at high risk for kidney disease, discuss NSAID administration with the primary care provider, and monitor fluid intake and urinary output. • Monitor patients for any changes in respiratory status and report even minor changes, such as elevated respiratory rate or decreased oxygen saturation, to the primary care provider, as they could be early signs of ACS. • Work with other members of the health care team to minimize disruptions to patients' dialysis schedule.
Psychosocial complications	• Assess patients, especially those with frequent ED visits and hospitalizations, for the presence of psychosocial health complications to identify any who may benefit from social work, psychiatric, or case management referral.

ACS = acute chest syndrome; AVN = avascular necrosis; NSAID = nonsteroidal antiinflammatory drug; SCA = sickle cell anemia; SCD = sickle cell disease; SCI = silent cerebral infarction; TCD = transcranial Doppler; VOE = vasoocclusive episode.

nurses can try to determine the type of chronic pain patients are experiencing. For example, a patient experiencing hip pain may require an orthopedic evaluation for AVN and possibly surgery. Nurses can also obtain a thorough medication history, including all opioid and nonopioid pain medications. Many patients with SCD require both short- and long-acting opioids and may use these medications for sleep or anxiety in addition to pain. Nurses can help patients understand how to use long-term opioids most effectively and to encourage the use of nonpharmacologic interventions as adjunctive therapy.

AVASCULAR NECROSIS

When the bone's blood supply is interrupted, bone tissue may die, a condition referred to as AVN. In people with SCD, AVN may result from VOEs in the bone's microcirculation that cause thrombosis, infarction, and ultimately, necrosis.[28] SCD severity and history of ACS may be risk factors for AVN,[2] which can occur in people of all ages and has been seen in children as young as five years.[29]

Although the VOEs of SCD can occur in any organ, they are especially common in the bone marrow, particularly in the femoral or humeral head, where they can cause unilateral or bilateral AVN.[28] In one study, the overall cumulative incidence of femoral head AVN was 15% by age 30; the median age at diagnosis was 27, and nearly a quarter of the patients underwent hip replacement surgery at a median age of 36.[2] Often, however, patients do not report the pain of AVN or are unable to obtain orthopedic follow-up. If undetected or untreated, AVN can cause permanent gait abnormalities and limb-length discrepancies, significantly impairing mobility.

Nursing implications. The comprehensive pain assessment conducted by the nurse may provide the first clues that the patient has AVN. If AVN is suspected and an orthopedic consultation has not yet been recommended, the nurse should discuss assessment findings with the primary care provider. If the patient is diagnosed with AVN, the nurse can help the patient and family understand the condition and the various procedures that may be considered to alleviate related pain and disability.

PRIAPISM

Priapism is an undesired, persistent, and often painful erection that can lead to erectile dysfunction, substantially impairing quality of life. In boys and men with SCD, priapism is a common complication. Researchers have calculated that the actuarial probability of male patients with SCA experiencing priapism was nearly 13% by age 10, more than 50% by age 15, and more than 89% by age 20.[30]

In a survey of 130 male patients with SCD being treated at one of five UK or Nigerian hospitals, a history of priapism was reported by 46 (35%) of respondents.[3] Within this group, 24 (52%) reported a history of severe, prolonged priapism (lasting more than 24 hours), which requires emergency treatment, and 33 (72%) reported a history of "stuttering" priapism—recurrent self-limited episodes, which are of much shorter duration. Stuttering episodes frequently heralded subsequent prolonged events, as only six of those reporting a history of severe priapism had no history of the stuttering type. In the group of 46 respondents, the mean age of priapism onset was 15, with 75% reporting their first episode before the age of 20 and 25% reporting a first episode before the age of 10. Sleep, sexual activity, and fever were frequent precipitating factors.[3]

Nursing implications. Prevention is key but is hindered by a lack of knowledge and awareness of the complication. In a survey of patients with SCA

NHLBI Expert Panel Recommendations for Managing Sickle Cell Disease[13, 15]

- In children with conditional (170 to 199 cm/sec) or elevated (> 200 cm/sec) transcranial Doppler (TCD) screening results, refer to a specialist with expertise in chronic transfusion therapy aimed at preventing stroke.
- In children and adults with sickle cell disease (SCD) and a vasoocclusive crisis associated with severe pain, rapidly initiate treatment with parenteral opioids.
- Treat avascular necrosis with analgesics and consult physical therapy and orthopedics for assessment and follow-up.
- Treat adults with sickle cell anemia who have three or more sickle cell–associated moderate-to-severe pain crises in a 12-month period with hydroxyurea.
- Regardless of clinical severity, to reduce SCD-related complications—such as pain, dactylitis, acute chest syndrome, or anemia—offer treatment with hydroxyurea to infants ages nine months or older, children, and adolescents who have sickle cell anemia.
- To ensure proper use of hydroxyurea and maximize benefits and safety, use an established prescribing and monitoring protocol.
- Use simple or exchange transfusion for children with TCD readings of > 200 cm/sec.

Facts About Sickle Cell Disease

- In the United States, the median age at death for men and women with sickle cell anemia is 42 and 48, respectively, and for men and women with HbSC, it is 60 and 68, respectively.[17]
- Among children with sickle cell disease (SCD), hospitalization and ED rates are estimated to be seven to 30 and two to six times higher, respectively, than those of same-age children without SCD.[10]
- Vasoocclusive episodes are the most common manifestation of SCD and the most common SCD-related ED diagnosis.[16]
- Aggressive incentive spirometry can reduce risk of acute chest syndrome,[18, 19] the primary cause of death[9] and one of the most frequent causes of hospitalization in both children and adults with SCD.[5]
- Strong social support is key to good disease management and quality of life. Patients lacking a support network may benefit from social work, psychiatric, or case management referral.

ages five to 20, only 7% of the men who had not experienced priapism were aware of what it was and knew it was a potential complication of SCA.[30] Patient teaching can include the use of analgesia, hydration, exercise, voiding, and warm baths to help prevent episodes.

Priapism can be an embarrassing problem for men with SCD, which may make it a difficult topic to discuss. Nurses may approach the discussion with an educational focus, providing answers to questions the patient may be afraid to ask. It's important for patients with SCD to be knowledgeable about the condition, since it has potentially devastating sequelae. Nurses can emphasize the importance of reporting recurrent events and seeking medical attention for prolonged episodes within four hours of onset.

ACUTE CHEST SYNDROME

ACS was the leading cause of death in the Dallas Newborn Cohort of patients with SCD[9] and one of the most frequent causes of hospitalization in SCD.[5] ACS is characterized by pleuritic chest pain, fever, rales on auscultation, and pulmonary infiltrates on chest X-ray.[5] It can resemble other acute respiratory illnesses, including pneumonia, asthma exacerbation, or bronchiolitis.

Patient age affects presentation, etiology, and severity of ACS. Incidence is highest in children ages two to four years,[5] but severity tends to be higher in adults, in whom studies have found mortality rates to be up to four times higher than in children.[31] Children are more likely to present with wheezing, fever, and cough, whereas adults more often present with dyspnea, pain (commonly in the chest, ribs, sternum, arms, or legs), productive cough, and hemoptysis. ACS frequently occurs during hospitalization for VOE and following surgery.[31, 32] In many cases, a cause is not evident or easily discernible. However, the most common etiologies are infection, followed by fat emboli from infarcted bone marrow. Following onset of symptoms, ACS can rapidly progress to respiratory failure, acute respiratory distress syndrome, or multisystem organ failure.

All patients with ACS should be hospitalized.[15] Analgesia should be sufficient to reduce respiratory splinting and hypoventilation without causing atelectasis.[33] Guidelines on the clinical management of ACS were published in 2015.[18] Among the recommendations in the guidelines are aggressive incentive spirometry (10 maximum inspirations) every two hours while the patient is awake to prevent atelectasis and the development of ACS during hospitalization for VOE.[18, 19] Although IV fluid administration is frequently a component of VOE management, excessive fluids can cause pulmonary edema and precipitate ACS.[18]

Nursing implications. Nurses can reduce the incidence of ACS by encouraging use of the incentive spirometer. Nurses can teach patients and family members the importance of appropriate use of the incentive spirometer and remain alert for subtle changes in respiratory status, such as increased work of breathing, decreased oxygen saturation, and new symptoms of cough or shortness of breath. Any change in respiratory status should be reported promptly to the primary care provider.

STROKE

Children and adults with SCD are at risk for cerebrovascular accidents such as ischemic stroke, hemorrhagic stroke, and silent stroke, also known as SCI. Analysis of clinical data on 4,082 SCD patients in the Cooperative Study of Sickle Cell Disease (CSSCD) provided the following prevalence estimates for cerebrovascular accidents in patients whose SCD genotype could be confirmed[34]:

- HbSS, 4.01%
- HbSβ^0-thalassemia, 2.43%
- HbSβ^+-thalassemia, 1.29%
- HbSC, 0.84%

SCIs, the most common strokes in children with SCD, are small infarctions detectable only by magnetic resonance imaging (MRI). Children with SCI typically present with no acute neurologic signs or symptoms. However, any child with SCD who presents with progressive neurocognitive deficits should

have MRI screening for SCI because, though SCI most commonly occurs in SCA (before age six in approximately 27% of these children and before age 14 in approximately 37%), it also occurs in 3% to 38% of patients with HbSβ+-thalassemia and in 5% to 31% of patients with HbSC.[6,35] Risk factors for SCI include low Hb levels, elevated systolic blood pressure, and male sex.[6]

Nursing implications. Nurses should maintain a high index of suspicion for SCI in patients with SCD who have neurologic signs or symptoms. Any acute changes in neurologic symptoms, including altered mental status during a hospitalization, should be immediately reported to the patient's primary care provider. Parents should be advised that poor academic performance is a red flag for potential neurocognitive deficits, possibly resulting from SCIs, and should be discussed with the child's health care team.

Ischemic stroke is common in both young children and adults with SCD. The CSSCD found that among patients with the HbSS genotype, in whom all types of cerebrovascular accidents are more prevalent, incidence of ischemic stroke is highest in children ages two to nine years.[34] In addition to having the HbSS genotype, risk factors for ischemic stroke include recent or recurrent ACS, elevated systolic blood pressure, low Hb level, prior transient ischemic attacks, and history of meningitis.

Transcranial Doppler (TCD) ultrasound. Prior to a first stroke, cerebral blood flow velocity may be elevated in children secondary to intracranial arterial stenosis.[36] Ischemia may also occur because the narrowing of the vessel reduces the amount of blood that can flow through to the tissues. Elevated blood flow velocity can be measured by noninvasive TCD ultrasound. Positive TCD screens may warrant blood transfusion or hydroxyurea to reduce blood flow velocity and prevent stroke.[37,38] The NHLBI recommends annual TCD screening of children with SCA, starting at age two and continuing through age 16, with referral to a specialist when results fall outside the normal range.[15]

Nursing implications. Nurses should reinforce with parents the need to seek care for any emerging neurologic symptoms in their children with SCD and teach them the importance of routine TCD screening of children with SCA.

Hemorrhagic stroke. The CSSCD found that hemorrhagic strokes occur most commonly in young adults ages 20 to 29 and were associated with a mortality rate of 24% overall and 26% in patients with the HbSS genotype.[34] Hemorrhagic strokes, which are associated with weakened cerebral blood vessels and aneurysms, usually occur suddenly, with symptoms of severe headache; vomiting; seizures; and changes in alertness, sensation, and language.[39] Risk factors for hemorrhagic stroke include low Hb level, high leukocyte count, and history of ACS.[34,39]

SPLENIC COMPLICATIONS

The spleen is one of the first organs affected by SCD. In children with SCA, hyposplenism, a physiological reduction in spleen function, is usually evident within the first year of life.[40] Splenic dysfunction is believed to stem from vasoocclusion of the organ by sickled erythrocytes and subsequent ischemia, fibrosis, and progressive atrophy of the organ. The spleen is a major filter of the blood and plays an important role in

Sickle Cell Disease Clinical Care Resources

American Society of Hematology
Pocket guides for clinicians can be downloaded or ordered, such as *Management of Acute Complications of Sickle Cell Disease*, *Health Maintenance and Management of Chronic Complications of Sickle Cell Disease*, and *Hydroxyurea and Transfusion Therapy for the Treatment of Sickle Cell Disease*.
www.hematology.org/Clinicians/Guidelines-Quality/Quick-Reference.aspx

Centers for Disease Control and Prevention
Materials and Multimedia on Sickle Cell Disease
https://www.cdc.gov/ncbddd/sicklecell/materials/index.html

National Heart, Lung, and Blood Institute
Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014
www.nhlbi.nih.gov/health-topics/evidence-based-management-sickle-cell-disease

National Institute for Children's Health Quality
On the Resources page, you can sign up for the Sickle Cell Disease Treatment Demonstration Program Compendium of Tools and Materials
www.nichq.org/resource/sickle-cell-disease-treatment-demonstration-program-compendium-tools-and-materials

immune defense as well as in vascular and blood homeostasis by removing senescent or altered red blood cells, microorganisms, and blood-borne antigens. Splenic damage puts patients at risk for immune dysfunction, vascular narrowing and occlusion, and severe bacterial infection. People with SCD remain at high risk for infection and sepsis throughout life.

Nursing implications. Patient and family teaching should emphasize the importance of receiving all recommended immunizations as well as annual influenza vaccines. Parents should be taught the importance of recognizing fever early and notifying their child's primary care provider. Elevated temperatures in hospitalized children should be immediately reported to the primary care provider because a sepsis evaluation may be necessary.

Acute splenic sequestration crisis. The splenic damage seen in patients with SCD is typically silent and progressive, but it becomes clinically apparent in acute splenic sequestration crisis (ASSC), a life-threatening complication involving rapid accumulation of sickled erythrocytes within the spleen.[41] ASSC is defined as sudden splenomegaly, a reduction in Hb concentration of at least 2 g/dL, and a normal or elevated basal reticulocyte count. The spleen enlarges within a period of hours, trapping a large portion of circulating erythrocytes and acutely worsening the anemia and circulatory failure. Symptoms include abdominal pain and distension, pallor, tachycardia, hypotension, and lethargy. Hypovolemic shock and death from cardiovascular collapse can occur within hours if untreated. In children with SCD, ASSC is one of the earliest life-threatening complications and may be the first clinical manifestation of the disease, occurring at a median age of 1.4 years, and as early as several weeks of age.[42] More than 12% of children with SCA experience ASSC, and 67% of those go on to have at least one more occurrence. In patients with HbSC, sequestration may occur in early and late adulthood.[43]

INFECTION AND SEPSIS

All SCD genotypes put patients at increased risk for severe infection—particularly invasive bacterial infection—throughout life. This increased susceptibility is largely due to splenic and immune dysfunction. Young children with SCA in particular are at elevated risk for pneumonia, septicemia, and meningitis.[44, 45] Incidence of these infections is lower in patients with HbSC and HbSβ$^+$-thalassemia genotypes because splenic function is typically normal or only minimally impaired in infancy. The risk, however, is present in later childhood and adulthood.[46] Other infections are also common in SCD. VOE of the bone and subsequent necrosis predisposes patients to osteomyelitis.[47] Parvovirus B19, which is often asymptomatic in those without SCD, can trigger aplastic crisis and life-threatening anemia in people with SCD.[48] Risk of catheter-related infections is also higher in SCD. Infection is a common precipitant of VOE and the most common cause of ACS.[32] Because of increased risks of infection, including infection with penicillin-resistant organisms, and incomplete vaccination in patients with SCD, any fever higher than 101.3°F (38.5°C) is considered a medical emergency requiring further evaluation.[15]

Nursing implications. Nurses should closely monitor vital sign trends, particularly temperature, in patients with SCD. Elevations in fever should be reported to the primary care provider immediately. Nurses should also teach patients and parents the importance of monitoring temperature and the need for age-appropriate immunizations.

ORGAN FAILURE

Renal disease. It's estimated that 16% to 18% of overall mortality in SCD is related to renal complications.[49] The most common renal complication in SCD is hyposthenuria (the inability to concentrate urine), which can lead to urinary frequency, enuresis, and increased risk of intravascular volume depletion.[50] Compared with the general U.S. population, patients with SCD are two to three times as likely to develop acute renal failure or chronic kidney disease.[51]

Adults with serum creatinine elevations above 1 mg/dL and children with elevations above 0.7 mg/dL are classified as having renal impairment and should receive nephrology consultation.[15] Microalbuminuria is the first manifestation of chronic kidney disease, and all patients with SCD should be screened for proteinuria annually beginning at age 10.[15] Risk factors for both acute renal failure and chronic kidney disease include the following[51]:

- advanced age
- male sex
- proteinuria
- diabetes
- hypertension
- chronic heart disease
- history of blood transfusion

Additional risk factors for chronic kidney disease include hypertension and dyslipidemia.[51]

Nursing implications. Nurses should be aware of the renal status of all patients with SCD and pay careful attention to kidney disease risk factors throughout hospitalization. Nurses should also maintain a high index of suspicion for proteinuria and decreased urine output, bearing in mind the patient's dialysis schedule. When patients are hospitalized, all health care team members need to work together to minimize disruptions to this schedule.

Multisystem organ failure in SCD is usually characterized by dysfunction of at least two or three major organ systems and often develops after several

days of hospital treatment for a VOE, when pain is improving.[22] Deterioration is typically rapid and associated with fever, nonfocal encephalopathy, and decreased Hb and platelet levels. Significant overlap between severe ACS due to fat emboli and multisystem organ failure has been noted. Multisystem organ failure is thought to result in part from diffuse microvascular occlusion and tissue ischemia.[4]

Nursing implications. Multisystem organ failure may occur rapidly in any patient with SCD. It is often signaled by changes in respiratory status or renal function. Nurses should consider any elevation in respiratory rate or reduction in oxygen saturation to be a potential early sign of ACS and report such signs to the patient's primary care provider. Renal function and IV fluid administration should also be monitored, especially in patients with a history of renal failure.

PSYCHOSOCIAL COMPLICATIONS

People with SCD suffer from a high incidence of social and behavioral health complications. Anxiety and depression have been reported to be as high as 7% and 28%, respectively, in these patients.[52] Social and behavioral health complications can occur throughout life.

Severe disease may cause young children to miss school, limit college and job opportunities for young adults, and reduce career opportunities for older adults. Such losses present significant financial challenges, often limiting access to health care, insurance, and necessary medications. Interviews with 147 ED patients with SCD, conducted by social workers over a 14-month period, revealed that 27% and 17% of patients, respectively, experienced difficulties obtaining insurance coverage and meeting copay requirements for prescriptions.[53] Other issues that may affect patients' ability to manage SCD effectively include navigating the time and transportation challenges involved in seeing numerous health care specialists while trying to work and take care of family responsibilities. It is not uncommon for patients to need to see an ophthalmologist, orthopedic surgeon, or nephrologist in addition to a sickle cell specialist and primary care provider.

Nursing implications. Nurses should assess patients, especially those with frequent ED visits and hospitalizations, for behavioral health complications to identify those who may benefit from social work, psychiatric, or case management referral. Patients should also be screened for depression, anxiety, and other social and behavioral determinants of health. When possible, nurses should refer patients with such complications to the social worker or case manager, who may be able to guide them in seeking extra support. A strong social support network is key to effective disease management and to a good quality of life in those with SCD. ▼

For five additional continuing nursing education activities related to sickle cell disease, go to www.nursingcenter.com/ce.

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